Management of Peripheral Nerve Sheath Tumors: 17-year Experience at the University Health Network

Daipayan Guha MD; Benjamin Davidson MD; Mustafa Nadi; Naif M. Alotaibi MD; Abhijit Guha MD, FRCSC, FACS; Gelareh Zadeh MD, PhD, FRCS(C)

Division of Neurosurgery, University of Toronto. Toronto, ON, Canada

Objective

We retrospectively review benign peripheral nerve sheath tumours (BPNST) managed surgically at the Toronto Western Hospital. Our primary objective was to identify independent predictors of neurologic deficit, tumour recurrence and extent of resection of BPNSTs.

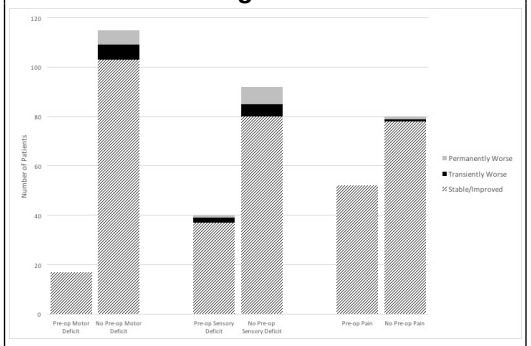
Background

PNSTs may be sporadic, as for most Schwannomas, or associated with autosomal-dominant neurofibromatoses (NF). 60% of NF1 patients develop neurofibromas, with a lifetime 5-15% risk of malignant transformation (1). 10% of NF1 patients develop primary MPNSTs; conversely, 50% of MPNSTs are associated with NF1. Known predictors of BPNST recurrence include subtotal resection, with some suggestion that neurofibromas, NF1+, and larger tumors are also predisposed to recur (2,3). Subtotal resection and prior office biopsy are identified predictors of neurologic deficit following BPNST resection (2,4).

Methods

All PNSTs treated surgically by the TWH Division of Neurosurgery from 1993-2010 were reviewed. Data was collected on patient age, gender, diagnosis of neurofibromatosis (NF), tumour histopathology, tumour location, tumour volume, and extent of resection. Postoperative motor, sensory and pain outcomes were dichotomized as stable/improved or worse than preoperative scores.

Figure 1



Postoperative motor, sensory and pain outcomes for 133 Schwannomas

Results

158 patients with 182 BPNSTs had adequate follow-up for analysis. Of these, 133 were Schwannomas, 21 of which were associated with a diagnosis of Schwannomatosis. There were 49 neurofibromas, 26 associated with NF1. Patients presenting with Schwannomas were significantly older than those with neurofibromas (Table 1). Patients with benign PNSTs presented typically with a painful mass and less frequently with motor deficits (Table 2).

Motor/Sensory/Pain Outcomes

New motor deficits were seen in 10.3% of Schwannomas (5.2% permanent) and 11.8% of neurofibromas (8.8% permanent); new sensory deficits were seen in 12.9% (7.5% permanent) and 3.7% (0% permanent), while new neuropathic pain was seen in 3.6% (1.8% permanent) and 16.7% (8.3% permaent), respectively (Figs. 1,2). The likelihood of worsened postoperative motor function was decreased in patients with fully resected tumours, or with preop deficits.

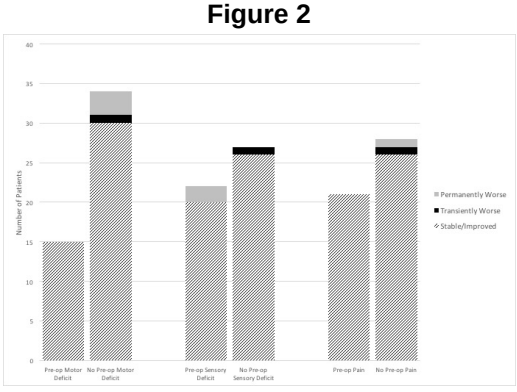
Extent of Resection

GTR was achieved for 76.7% of Schwannomas and 44.9% of neurofibromas (51.4% of non-plexiform neurofibromas). The only independent predictor of STR was tumour type (for neurofibromas, OR of GTR =0.25, p<0.001). Tumour location was predictive of extent of resection for Schwannomas only: extremity tumours were more readily resected than plexal tumours, and those in the brachial plexus were more readily resected than those in the lumbosacral plexus.

Tumour Recurrence

Recurrence rates for Schwannomas and neurofibromas in our cohort were 5.3% and 8.2%, respectively. Recurrence of Schwannomas and neurofibromas was seen more frequently in patients diagnosed with NF3 and NF1, respectively; subtotal resection was associated with increased recurrence for all benign lesions (OR=13.16, p=0.007).

Figure 2



Postoperative motor, sensory and pain outcomes for 49 neurofibromas

Table 1

Characteristic	Value
Age (years)	45.2 ± 15.8
Schwannoma	48.4 ± 15.6
Neurofibroma	36.6 ± 12.8
Gender (M:F)	96:79
Schwannoma	64:51
Neurofibroma	22:21
NF status	
NF1	37
NF2	3
NF3	8
Tumour histopathology	
Schwannoma	115
Neurofibroma	43
Mean follow-up (months)	29.5 (range 1-302)

Patient demographics

Table 2

Tumour Type	Weakness	Numbness/ Paresthesiae	Tinel	Pain	Mass Only
Schwannoma	17 (12.8%)	40 (30.1%)	86 (64.7%)	77 (57.9%)	30 (22.6%)
Neurofibroma	15 (30.6%)	22 (44.9%)	25 (51.0%)	37 (75.5%)	2 (4.1%)

Symptomatic presentation of 182 BPNSTs

Conclusions

- Extent of resection is the primary independent predictor of motor outcome
- GTR is achieved less often for neurofibromas than Schwannomas
- Extent of resection and NF status are independent predictors of recurrence

References

1) Tucker et al. Association between benign and malignant peripherla nerve sheath tumors in NF1. Neurology 2005.
2) Montano et al. Tumors of the peripheral nervous system: analysis of prognostic factors in a series with long-term follow-up and review of the literature. JNS 2015.
3) Safaee et al. Association of tumor location, extent of resection, and neurofibromatosis status with clinical outcomes for 221 spinal nerve sheath tumors. Neurosurgical Focus 2015.
4) Levi et al. The surgical management of symptomatic peripheral nerve sheath tumors. Neurosurgery 2010.